

Case Report

Endolymphatic sac tumor of the left fossae temporalis and fossa cranii posterior: report of a rare case

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Abstract: Endolymphatic sac tumor (ELST) is a rare low-grade non-metastasizing adenocarcinoma of endolymphatic sac origin that arises in the petrous part of the temporal bone. Here we present a case of endolymphatic sac tumor of the head in a 53-year-old man who had symptoms in his left ear since 40 years ago after drowning and found a mass about 2 cm around his left ear 8 years ago, which grows slowly to about 10 cm in appearance. MRI detected a mass about 8.4 cm×6.2 cm, located in the left fossae temporalis and fossa cranii posterior, with the left petrous bone in the center of the tumor. Histologically, the tumor cells are low cuboidal cells without marked atypia and show papillary architecture and glandular formation. Immunohistochemical staining shows that the tumor cells were positive for CK (AE1/AE3), but negative for vimentin, Pax-8, TTF-1 and PSA. Ki67 index was lower than 5%.

Keywords: Endolymphatic sac tumor, fossae temporalis, fossa cranii posterior, petrous bone

Background

Endolymphatic sac tumor (ELST) is a rare non-metastasizing adenocarcinoma of endolymphatic sac origin, which grows slowly and widely invades the petrous bone [1]. The ages of the patients range from 4 to 85 years old without notable age and gender preference [2]. In this case, the 53-year-old patient had symptoms of his left ear after drowning 40 years ago and found a mass around the ear 8 years ago. The mass grows slowly and turns to about 10 cm in appearance and at that time the patient came to the clinic of our hospital without any treatment of the mass before. In most cases, ELST cells are cuboidal cells and form a variable papillary-glandular architecture. Thus a variety of primary and metastatic papillary tumors should be differentiated from this tumor. MRI imaging of the patient shows a huge mass which located in the left fossae temporalis and fossa cranii posterior, around the left petrous bone, which destructed most of the temporal and occipital bone indicating it a locally invasive tumor. Though aggressive papillary tumor of the middle ear also shows locally invasive

behavior, it has been suggested that the widespread involvement of the surrounding organs including temporal bone indicates the tumor may arise from a primary papillary adenocarcinoma of the endo-lymphatic sac [1]. In addition, current evidence suggests that so called “aggressive papillary middle ear tumor” is ELST with extension into the middle ear [1]. As the tumor extended over a wide expanse of space and destructed large areas including temporal and occipital bone, and the bone structures of the labyrinth, internal auditory canal and external auditory canal, the tumor should be considered as ELST rather than aggressive papillary tumor of the middle ear. The immunohistochemical staining pattern of the tumor with negative staining of Pax-8, TTF-1 and PSA excluded the diagnosis of metastatic papillary tumor from thyroid or prostate.

Case presentation

Clinical history

A 53-year-old man referred to our hospital for a bulging about 10 cm around his left ear. He

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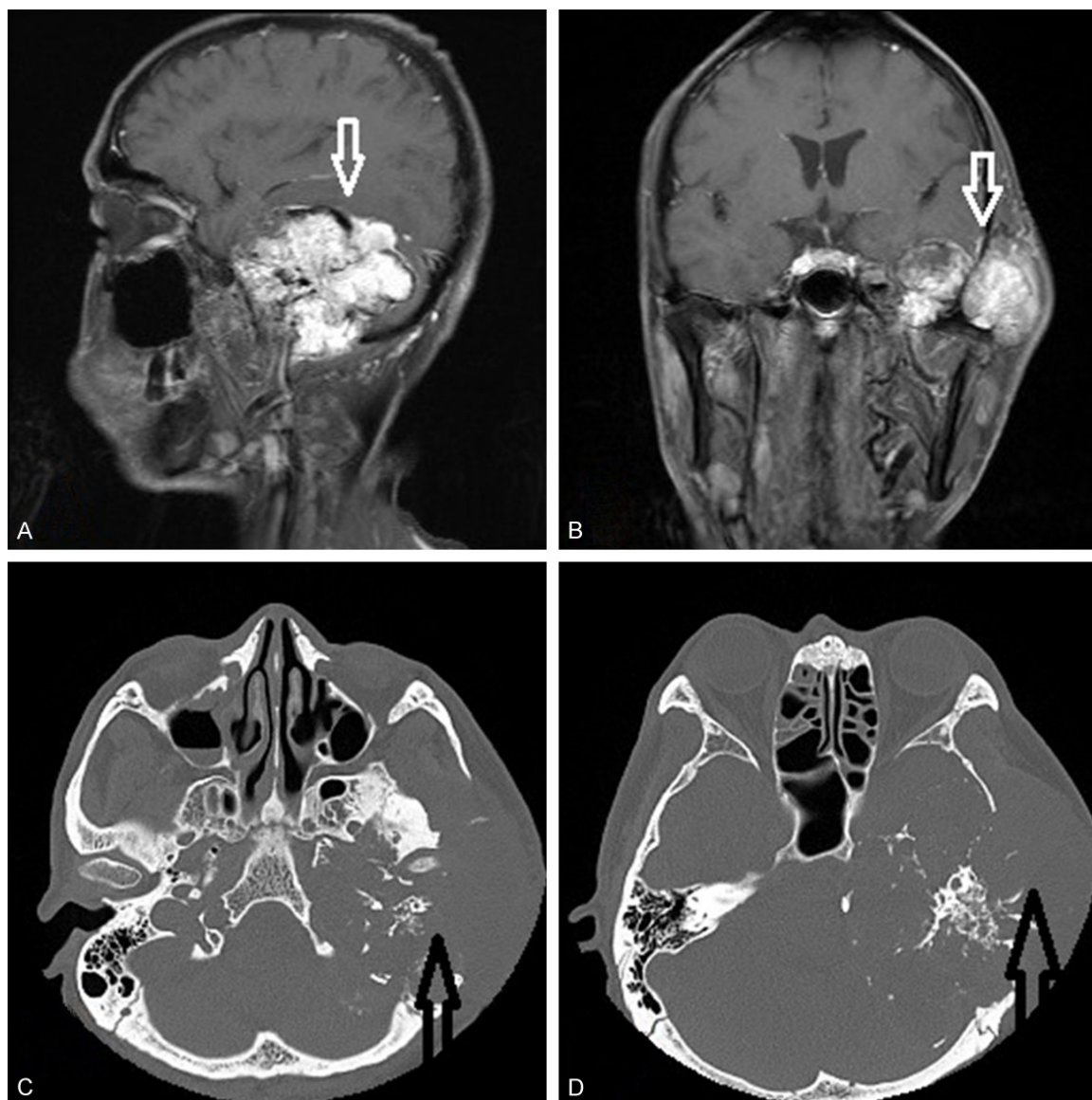


Figure 1. Imaging of the tumor. MRI (A, B) and CT (C, D) detected a huge mass (the arrows) about 8.7 cm×8.4 cm×6.2 cm (transverse diameter, anteroposterior diameter, vertical diameter) in the left fossae temporalis and fossa cranii posterior, with the left petrous bone in the center of the tumor. The temporal and occipital bone and the bone structures of the labyrinth, internal auditory canal and external auditory canal were vastly destroyed (A-D).

found the mass 8 years ago and at that time the mass was about 2 cm and he didn't go to hospital to receive special treatment for the mass. The patient has experienced a drowning accident 40 years ago and after that he had repeated symptoms in his left ear including bleeding and purulence. The patient suffered from facial paralysis and deafness 2 years after the drowning. The patient received surgery 2 years after in another hospital, but the details of the procedure and the pathological diagnosis were not very clear based on the oral

account of the patient. He received another surgery for bleeding and purulence in his left ear 4 years after the first surgery. The patient accidentally found a mass about 2 cm behind his left ear 8 years ago. He did not go to hospital to take special treatment for the mass. According to the patient, the mass grows gradually 4 years ago after he had a car accident and it became about 10 cm large in appearance, bulging around the ear when he came to the clinic of our hospital. During the period, the patient also had repeated bleeding and puru-

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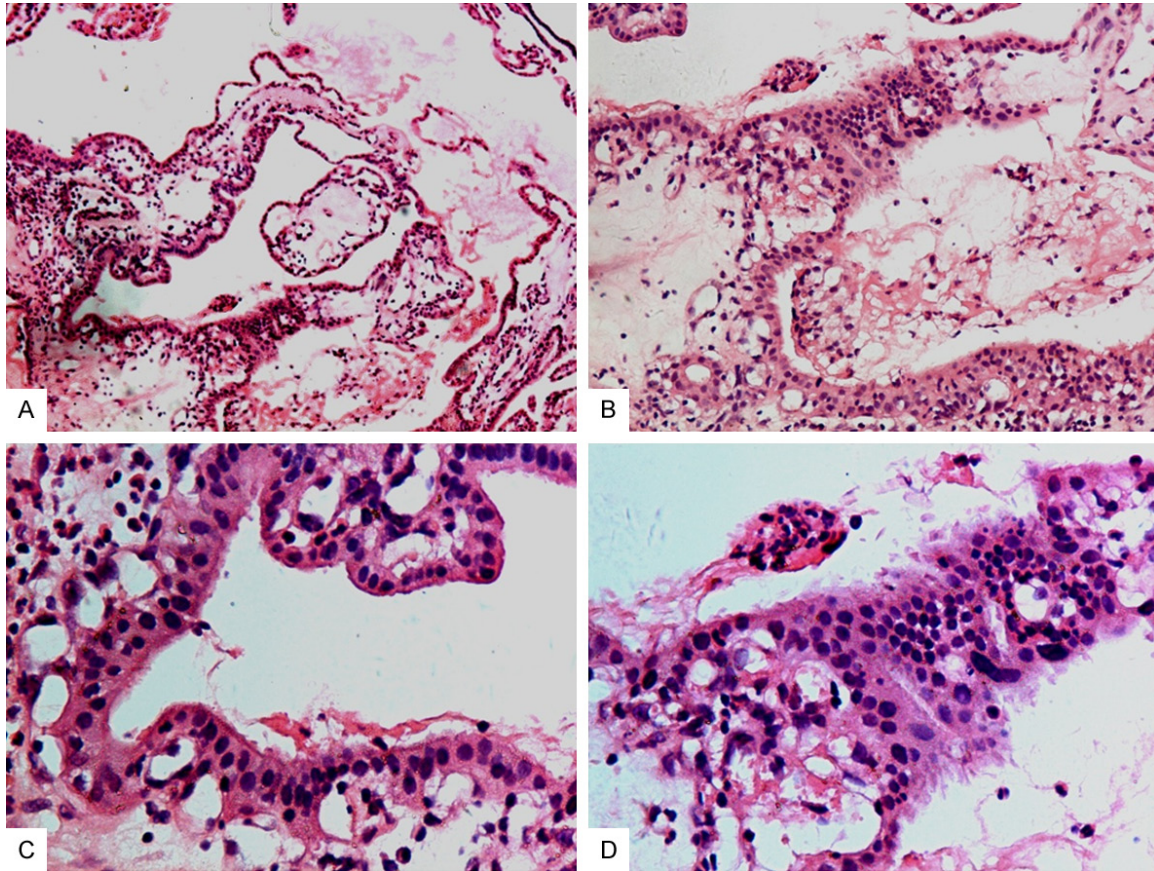


Figure 2. Microscopic findings of the tumor. The tumor cells show epithelial features including papillary architecture and glandular formation (A, B, $\times 200$). Most of the tumor cells are cubic or short-columned in shape without marked atypia (C, D, $\times 400$). The cytoplasm of the cells is eosinophilic and abundant (C, D, $\times 400$). The nuclei are round and the karyoplasmic ratio is small. The mitotic activity was low (C, D, $\times 400$).

lence in his left ear. No ulcer and purulence was found in the skin around the ear. The patient had no fever.

Materials and methods

Specimens were embedded in paraffin blocks and cut into 4 μm -thick sections. The sections were dewaxed in xylene and rehydrated stepwise in descending ethanol series. Endogenous peroxidase activity and non-specific binding were blocked using 3% H_2O_2 and non-immune sera. The sections were incubated with the following primary antibodies: AE1/AE3 (1:50, DAKO), Ki67 (1:200, DAKO), Pax-8 (1:50, DAKO), PSA (1:100, DAKO), TTF-1 (1:100, DAKO), and vimentin (1:200, DAKO) overnight at 4°C. The procedure of the catalyzed signal amplification system (Maixin Biotechnology, Fuzhou, Fujian, China) was according to the manufacturer's instructions. A standard avidin-biotin complex

method with biotinylated secondary antibodies (Maixin) was used for detecting the antibodies.

Results

Imaging and gross features

MRI and CT detected a huge mass about 8.7 cm \times 8.4 cm \times 6.2 cm (transverse diameter, anteroposterior diameter, vertical diameter) (**Figure 1A-D**) in the left fossae temporalis and fossa cranii posterior, with the left petrous bone in the center of the tumor. The lower margin of the mass was down to C1 and the top rim was up to the quadrigeminal cistern level (**Figure 1C, 1D**). The surface configuration of the tumors was multiply lobulated (**Figure 1A, 1B**). The boundary between the mass and the surrounding brain tissue is still clear. But the left cerebellar and brainstem was compressed and moved right. The left temporal lobe of the

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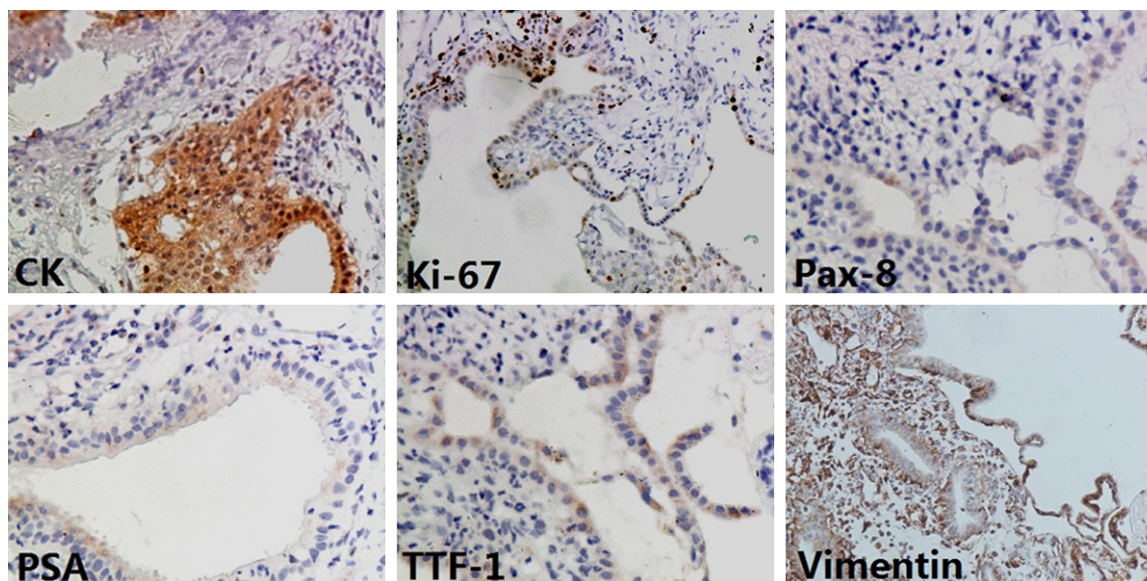


Figure 3. Immunohistochemical examination. The tumor cells were positive for CK (AE1/AE3) and negative for vimentin, TTF-1, PAX-8 and PSA. Ki67 index was lower than 5%.

brain was also compressed and moved up (**Figure 1A, 1B**). The temporal and occipital bone and the bone structures of the labyrinth, internal auditory canal and external auditory canal were vastly destroyed (**Figure 1A-D**). The left jugular foramen and facial nerve root were not clearly displayed (**Figure 1A-D**). The structure of the right side of the auditory canal was entire. The mucosa of bilateral ethmoid sinus and the right maxillary sinus were thickening (**Figure 1A, 1B**). The samples of the mass inspected are obtained by needle aspiration biopsy. Because the patient didn't receive surgery eventually we can observe the overall appearance of the tumor mainly by MRI and CT imaging described as above.

Microscopic features

Histologically, the tumor cells show epithelial features, such as papillary architecture and glandular formation (**Figure 2A, 2B**). Most of the tumor cells are cubic or short-columned in shape without marked atypia (**Figure 2C, 2D**). The cytoplasm of the cells is eosinophilic and abundant. The nuclei are round and the karyoplasmic ratio is small (**Figure 2C, 2D**). The mitotic activity was low (**Figure 2C, 2D**). Some areas show active proliferation of tumor cells with pseudostratified architecture (**Figure 2D**).

Immunophenotype

The tumor cells were positive for CK (AE1/AE3) and negative for vimentin, TTF-1, PAX-8 and PSA (**Figure 3**). Ki67 index was lower than 5% (**Figure 3**).

Discussion

ELSTs are rare non-metastazing aggressive papillary lesions arising from the intrapetrous portion of the endolymphatic sac [1-9]. However, there are still some different opinions on the origin of this tumor [1, 10, 11]. There is another tumor termed aggressive papillary middle ear tumor which has similar histological appearances as ELSTs [1]. However current evidence suggests that the so-called "aggressive papillary middle ear tumor" is ELST with extension into the middle ear, though an origin in the middle ear in some cases of this neoplasm has not been definitely excluded [1].

Here we present a case of ELST in a 53-year-old man who has a long history of left ear illness. He had symptoms of ear including bleeding and purulence since 40 years ago after a drowning accident. He found a mass behind his left ear 8 years ago, which became a huge mass and was biopsied in the clinic of our hospital. Though some of these tumors are related to von Hippel-Lindau disease (VHL) [12-16], our case also

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indicates a possible carcinogenesis mechanism related to long-term stimulation by inflammation.

Histologically, the tumor cells formed papillary and glandular architectures [1]. Thus it should be differentiated with primary and metastatic tumors with similar histological features such as the so called "aggressive papillary middle ear tumor", thyroid papillary carcinoma and prostatic carcinoma. The MRI and CT imaging revealed a huge mass which destructed most of the temporal and occipital bone, and the bone structures of the labyrinth, internal auditory canal and external auditory canal. As mentioned above, according to the imaging findings, it should be considered ELST rather than aggressive papillary middle ear tumor. The immunohistochemical staining shows that the tumor cells were negative for PSA, PAX-8 and TTF-1, which does not support the diagnosis of metastatic tumor of thyroid papillary carcinoma and prostatic carcinoma. What is more important is that the patient didn't show any clinical symptoms of these tumors.

Conclusion

ELST is a rare low-grade adenocarcinoma of endolymphatic sac origin. The tumor usually invades surrounding organs including bones and the ear canal, though histologically, the tumor cells show no marked cell atypia. Imaging findings are important to understand the site and the growth pattern of the tumor and are dispensable for the diagnosis. Several metastatic tumors including prostatic carcinoma and thyroid papillary carcinoma should be excluded.

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Disclosure of conflict of interest

None.

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